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CASE REPORT

A Unique Case of Osteitis Fibrosa Cystica with Postoperative Hungry Bone Syndrome and Hypocalcemic Cardiac Failure

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ABSTRACT

Osteitis fibrosa cystica, a manifestation of severe hyperparathyroidism presenting with crippling bone deformities is a rare presentation these days. We report a case of 40-year-old male patient who presented with generalized aches and pains and bony deformities. Radiographs showed diffuse osteopenia, brown tumors and pathological fractures of phalanges but common manifestations associated with hyperparathyroidism like pancreatic calcifications and nephrolithiasis were absent. Serum calcium and parathyroid hormone levels were elevated while ultrasound imaging of neck showed the presence of a left lower parathyroid adenoma and was confirmed by Tc99-sestamibi scan. Large parathyroid lesion along with high calcium levels and severely elevated PTH puts this patient in high-risk category for postoperative hungry bone syndrome leading to severe hypocalcemia postoperatively. Hypocalcemia usually results in neuromuscular irritability manifesting as paresthesia, Chvostek and Trousseau sign, carpopedal spasm and even convulsions in severe cases. However, our patient had none of the common manifestations, but developed hypocalcemic cardiac failure postoperatively. Case history and management of case is presented.

Keywords: Hyperparathyroidism, Parathyroid adenoma, Hypocalcemia, Hypocalcemic cardiac failure, Hungry bone syndrome.

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INTRODUCTION

'Osteitis fibrosa cystica' the term used to describe advanced skeletal manifestations of hyperparathyroidism, was first coined by Von Recklinghausen in 1891[1] and is characterized by fractures, deformities, bone cysts and brown tumors.[2] Such advanced skeletal presentations are rare in the current era due to early detection of serum calcium abnormalities by automated analyzers.

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We report a case of parathyroid adenoma presenting selectively with osteitis fibrosa cystica without any other features of hyperparathyroidism. Postparathyroidectomy patient developed hungry bone syndrome, hypocalcemic cardiac failure and required massive doses of calcium for maintenance of serum calcium levels.

CASE REPORT

A 40-year-old male presented with 2 years history of generalized bone pains and deformity of spine. He had difficulties in walking and lost 25 kg weight within the span of 1 year. On examination, patient was emaciated and restricted to bed due to severe pains and asthenia. Patient had gross bony abnormalities like kyphoscoliosis, pectus excavatum and deformed phalanges. There was palpable mass of 3 × 4 cm on the left side of neck. The mass was firm in consistency and moved with deglutition.

Skeletal survey revealed generalized osteopenia involving skull, long bones, phalanges and severe deformity of rib cage and vertebral column (Figs 1 to 4). Brown tumors and pathological fractures of phalanges were present. Serum biochemistry showed elevated serum calcium (12.7 mg/dl), elevated parathyroid hormone (PTH) (1886 pg/ml), elevated serum alkaline phosphate levels (3815 IU/ml) and low serum 25 (OH) D3 levels (9 pg/dl). High resolution ultrasonography of neck showed a hypoechoic lesion of approximately 3.6 × 3 × 2 cm in size located inferolateral to the left lobe of thyroid. Tc 99-sestamibi scan suggested a hyper functioning left lower parathyroid adenoma. Imaging the pancreas and urinary tract showed no abnormalities. Preoperative evaluation revealed a normal cardiac function (electrocardiogram and echocardiography).

A diagnosis of parathyroid adenoma was made and it was also recognized that patient had high-risk case features for postoperative hungry bone syndrome (high serum calcium and serum PTH levels, severe osteopenia, and large parathyroid gland).[3] Vitamin D deficiency was corrected preoperatively as the patient had severe deficiency which may have contributed to postoperative hypocalcemia. Patient was a high-risk case for general anesthesia because of severe kyphoscoliosis compromising his pulmonary function and poor performance

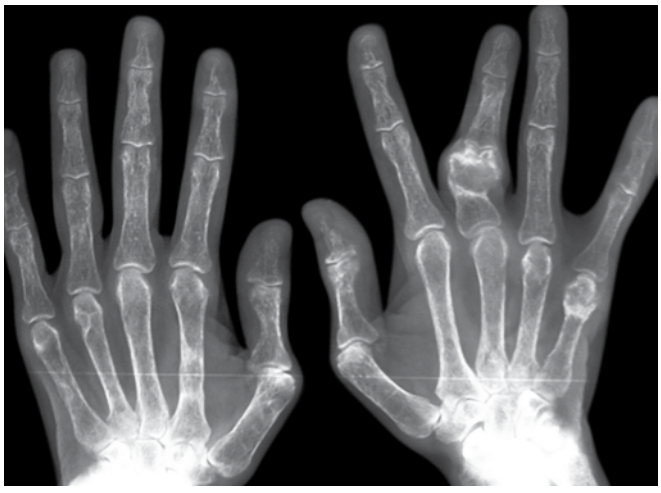


Fig. 1: Radiograph of hand showing tuft resorption of distal phalanges, subperiosteal resorption and brown tumors

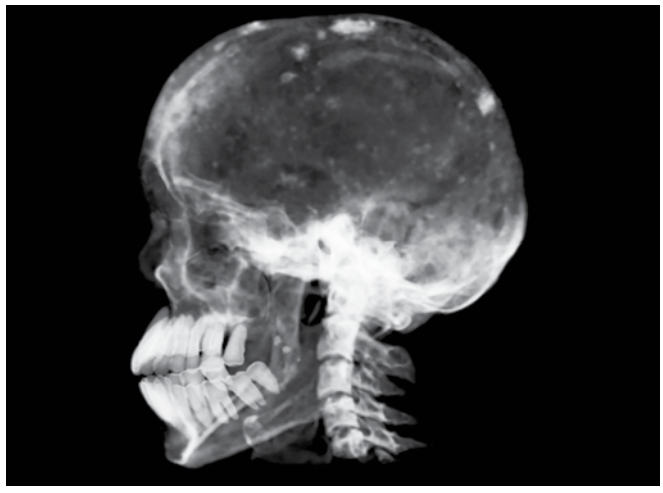


Fig. 2: Radiograph of skull showing moth-eaten appearance

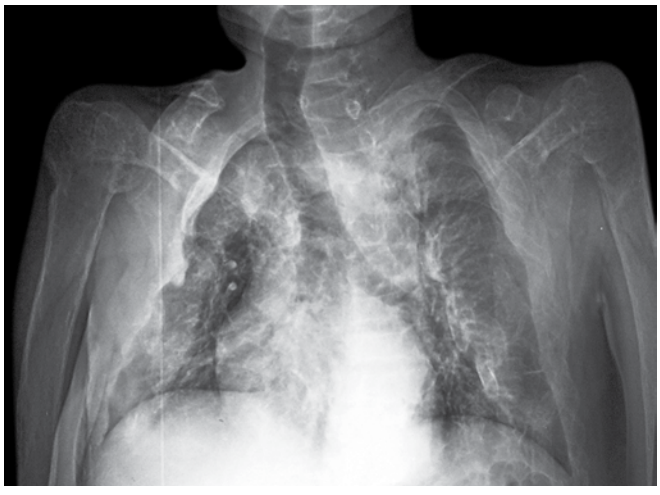


Fig. 3: Radiograph of chest showing severe osteopenia and deformity. Note the gross deviation of trachea

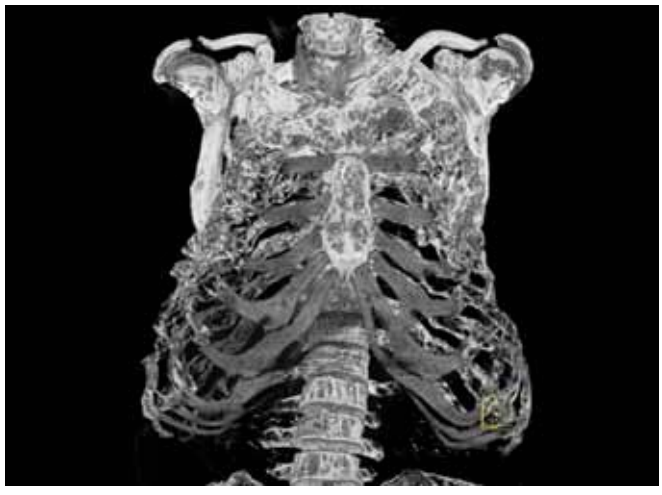


Fig. 4: Three-dimensional reconstruction of rib cage showing severe osteoporosis and deformities

status. Tracheal deviation added to patient's anesthetic risk. Intraoperatively, the thinned out left lobe of thyroid was found stretched over the parathyroid swelling (Fig. 5). Hence, left lobe of thyroid was excised along with enlarged left lower parathyroid gland and postoperative histopathological examination confirmed the presence of a large parathyroid adenoma. Patient was ventilated for 24 hours postoperative in view of his risk of tracheomalacia from his deformed and deviated trachea.

Immediate postoperative PTH value was 23 pg/ml. Calcium levels fell to 6 mg/dl within 3 hours after surgery. Postoperatively, the patient was maintained on continuous calcium infusion through a subclavian central line. Infusion was supplemented with periodic boluses of 10% calcium gluconate fourth hourly based on repeated serum calcium measurements.

In spite of aggressive calcium supplementation in the postoperative period, the patient developed congestive cardiac failure within 8 hours of surgery. There was steady decrease of blood pressure, followed by decreased urine output, raised CVP and lung crepitations. Emergency echocardiography confirmed the diagnosis. Inotropic support with noradrenaline and dopamine was started which improved blood pressure and urine output. The need for pressor support weaned on fifth postoperative day when serum calcium levels reached 9 gm/dl.

In spite of severe hypocalcemia, as low as 6 mg/dl (preoperative level of 12.7 mg/dl) patient never developed Chvostek sign, carpopedal spasms or other usual manifestations of neuromuscular irritability except for the rare manifestation of cardiac failure.

Hypocalcemia (<8.4 mg/dl) lasted beyond 4th postoperative day allowed us to diagnose this as hungry bone syndrome. It was successfully managed with calcium supplementation initially with combination of intravenous and oral routes (Graphs 1 and 2).

DISCUSSION

Primary hyperparathyroidism (PHPT) is characterized by hypercalcemia due to autonomous secretion of PTH.

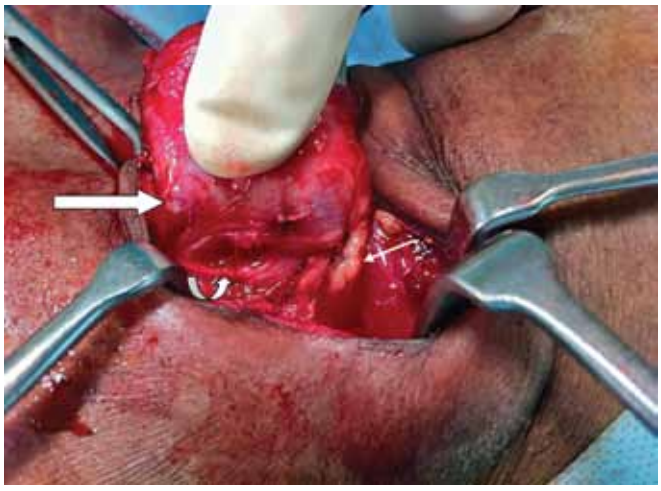


Fig. 5: Intraoperative photograph showing parathyroid adenoma arising from lower parathyroid (bold straight arrow). Also note the recurrent laryngeal nerve (bold curved arrow) and superior parathyroid (straight arrow)

Sporadic parathyroid adenoma is the most common cause accounting for 80% of cases, followed by parathyroid hyperplasia (sporadic and inherited) which is the cause in 15 to 20% of cases. Parathyroid carcinoma accounts for less than 1% of cases.

'Painful bones, renal stones, abdominal groans and psychic moans' is the mnemonic classically used to summarize the clinical features of hyperparathyroidism. Apart from bone changes, it includes renal stones, pancreatitis leading to abdominal pain and neuropsychiatric symptoms.[4]

The introduction of multichannel automated analyzers for biochemical screening in the 1970s has facilitated the early detection of PHPT. Currently in the western world, PHPT is a relatively common disease with an incidence of 1% in adults, majority of patients are diagnosed with asymptomatic hypercalcemia and presentation with classical symptoms and signs is rare.[5] In contrast in developing countries like India, PHPT remains a rare disease with advanced presentations.

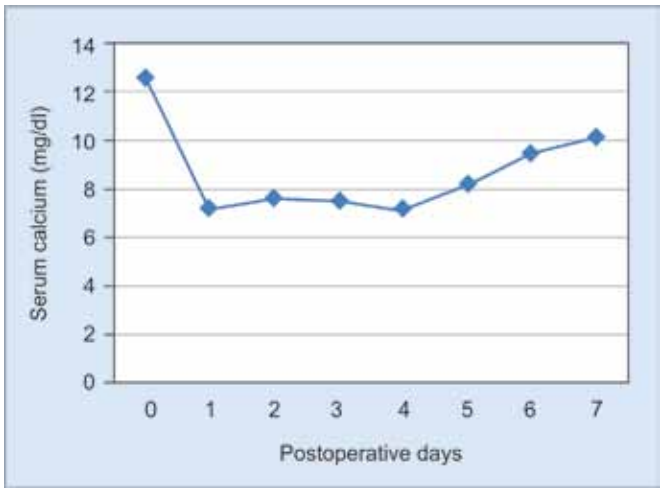
The diagnosis of PHPT in our case of hypercalcemia was relatively straightforward because of elevated serum parathormone and serum calcium. Familial hypercalcemic hypocalciuria is the only other condition that can present with a combination of elevated serum calcium and parathyroid hormone. Familial hypercalcemic hypocalciuria presents at a younger age with hypercalcemia and low urine calcium excretion (usually < 100 mg/24 hr). Positive family history is usually present.

Chronic elevation of PTH stimulates osteoclastic activity and inhibits bone formation thereby mobilizing calcium from bone causing extensive bone resorption. This process is responsible for osteopenia, peritrabecular fibrosis, formation of cysts and brown tumors ultimately.[6]

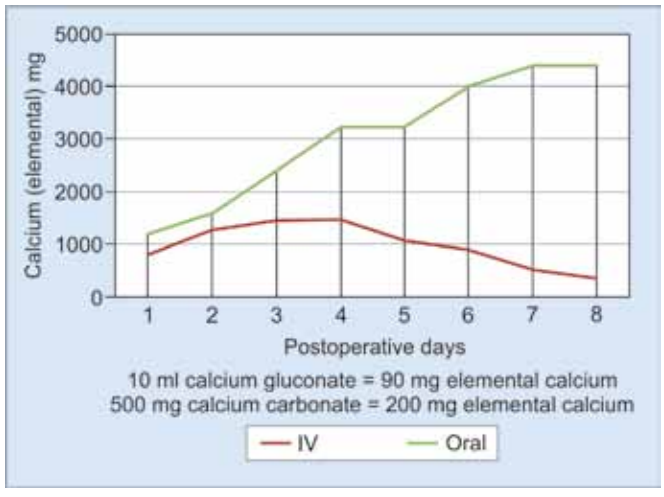
Our patient had extensive osteopenia with bone pains as the initial presenting picture. An interesting feature of this patient was in spite of extensive osteopenia, he escaped major fractures of weight-bearing bones probably due to extensive muscle weakness and fatigue confining him to bed. However, brown tumors and pathological fractures of phalanges were present.

Severe vitamin D deficiency in this patient might be a contributing factor for severe bone disease in this patient. Several studies have suggested that vitamin D deficiency in patients PHPT is associated with more severe bone disease, larger tumors, higher PTH levels and increased postoperative complications including hungry bone syndrome.[7-9]

Postoperative hungry bone syndrome and cardiac failure are other unique features of this case. The diagnosis of HBS is considered when patients have persistent hypocalcemia (< 8.4 mg/dl) after 3 to 4 days of parathyroidectomy. Large amounts of oral calcium (up to 12 gm/day) is required to manage serum calcium levels. Intravenous calcium infusion is administered in acute phase until symptoms of tetany recover and oral administration can maintain adequate levels.[10]



Graph 1: Postoperative serum calcium levels



Graph 2: Postoperative calcium supplementation

Our patient had rapid onset of hypocalcemia and cardiac failure that occurred within 8 hours of parathyroidectomy. Patient did not manifest any of the common features of tetany even when serum calcium levels were as low as 6.0 mg/dl. Literature search revealed few case reports of hypocalcemic cardiac failure but to our knowledge there are no reports of hypocalcemic cardiac failure exclusive of other signs and symptoms of hypocalcemia in acute setting.[11-14]

In this patient, the diagnosis of hypocalcemic cardiac failure is strongly suggested by the following facts:

- Patient had normal preoperative cardiac function and heart failure developed only after a fall in serum calcium postoperatively.
- Normalization of serum calcium resulted in prompt recovery from cardiac failure indicated by decreased need for pressors and improved urine output.
- Electrocardiogram, echocardiography and cardiac enzymes were not suggestive of myocardial ischemia.

It is known that hypocalcemia prolongs the duration of second phase of the action potential of cardiac muscle. The concentration of the extracellular Ca^{++} ion is thought to have a direct effect on the strength of the myocardial contraction through process of excitation-contraction coupling.[15] Hypomagnesemia has also been linked to reduce cardiac contractility.[16] Our patient had normal magnesium levels throughout the postoperative period which rules out any effects of hypomagnesemia on heart.

CONCLUSION

Severe hypocalcemic cardiac failure is a possible life-threatening complication even in the absence of classic cues to postoperative hypocalcemia. Hence, close monitoring of patients with high-risk features of hungry bone syndrome and aggressive correction of hypocalcemia is required to prevent complications.

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